Surgical Management of Moyamoya Syndrome

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ABSTRACT

Moyamoya syndrome, a vasculopathy characterized by chronic progressive stenosis at the apices of the intracranial internal carotid arteries, is an increasingly recognized entity which is associated with cerebral ischemia. Diagnosis is made on the basis of clinical and radiographic findings, including a characteristic stenosis of the internal carotid arteries in conjunction with abundant collateral vessel development. Adult moyamoya patients often present with hemorrhage, leading to rapid diagnosis. In contrast, children usually present with transient ischemic attacks or strokes, which may prove more difficult to diagnose because of patient's inadequate verbal and other skills, leading to delayed recognition of the underlying moyamoya. The progression of disease can be slow, with rare, intermittent events, or it can be fulminant, with rapid neurologic decline. However, regardless of the course, it is apparent that moyamoya syndrome, both in terms of arteriopathy and clinical symptoms, inevitably progresses in untreated patients.

Surgery is generally recommended for the treatment of patients with recurrent or progressive cerebral ischemic events and associated reduced cerebral perfusion reserve. Many different operative techniques have been described, all with the main goal of preventing further ischemic injury by increasing collateral blood flow to hypoperfused areas of the cortex, using the external carotid circulation as a donor supply. This article discusses the various treatment approaches, with an emphasis on the use of pial synangiosis, a method of indirect revascularization. The use of pial synangiosis is a safe, effective, and durable method of cerebral revascularization in moyamoya syndrome and should be considered as a primary treatment for moyamoya, especially in the pediatric population.

KEYWORDS: Moyamoya, pial synangiosis, ischemia, stroke

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Moyamoya syndrome is a vasculopathy characterized by chronic progressive stenosis to occlusion at the apices of the intracranial internal carotid arteries, including the proximal anterior cerebral arteries and middle cerebral arteries. It has been associated with ~6% of childhood strokes.[1,2] This progressive stenosis occurs simultaneously as characteristic arterial collateral vessels develop at the base of the brain. These collateral vessels, when visualized on angiography, have been likened to the appearance of haze, a cloud or a puff of smoke—which translates to "moyamoya" in Japanese. This disease was first described in the Japanese literature in 1957 when Takeuchi and Shimizu reported a case of "hypoplasia of the bilateral internal carotid arteries," but was first named as moyamoya 1969 by Suzuki and Takaku.[3,4]

EPIDEMIOLOGY

First described in Japan, moyamoya syndrome has now been observed throughout the world.[5] Although considered until recently as more prevalent in the Asian population, it can also affect individuals of many ethnic backgrounds, and there is an increasing awareness of this disease in the Americas and in European populations.[6] In Japan, it is the most common pediatric cerebrovascular disease, affecting females almost twice as often as males.[1] In Europe, a recent study cited an incidence of 0.3 patients per center per year, which is approximately one tenth of the incidence in Japan.[7] In the United States and Korea, reports corroborated historical claims of a bimodal age distribution of moyamoya, one group in the pediatric age range (around the first decade of life) and a second group of adults in the 30- to 40-year-old range. Both groups found that children were more likely to present with ischemic events (either strokes or transient ischemic attacks (TIAs)) than hemorrhage, more common in the adult group.[8,9] The goal of therapy is to arrest or reverse the course of these ischemic events.

CLINICAL PRESENTATION

Adult moyamoya patients often present with hemorrhage, leading to rapid diagnosis. In contrast, children usually present with TIAs or strokes, which may prove more difficult to diagnose because of these patients' inadequate verbal and other skills, leading to delayed recognition of the underlying moyamoya.[10] TIAs may be precipitated by events common in children, such as hyperventilation with crying; the hyperventilation can induce cerebral vasoconstriction in the vessels of these patients that have normal autoregulation and that are maximally dilated because of the patients' chronic ischemia. The subsequent constriction of these previously dilated vessels can in turn cause a significant reduction in the already marginal cerebral perfusion in these patients and lead to TIA or stroke.

Headache is a common presenting symptom in children with moyamoya. Although the etiology is unclear, there is speculation that dilatation of meningeal and leptomeningeal collateral vessels may stimulate dural nociceptors. Seizures (both focal and global), focal neurologic deficits, choreoathetotic movements, and hemorrhage are also presenting signs and symptoms of these patients. Table 1 summarizes the clinical presentation in a recently published series of 143 patients from our institution.[10a] Lastly, a rare but reported manifestation of patients with moyamoya syndrome is

Table 1 Symptoms at Presentation in 143* Patients

Symptom	Number	Percent
Stroke	97	67.8%
TIAs (including drop attacks)	62	43.4%
Seizures	9	6.3%
Headache	9	6.3%
Choreiform movements	6	4.2%
Incidental	6	4.2%
Intraventricular or intracerebral bleed	4	2.8%

*Symptom totals are greater than patient numbers, since some patients had multiple symptoms at presentation.
TIA, transient ischemic attacks.

nontraumatic acute subdural hematoma, which is presumed to result from spontaneous rupture of transdural collateral vessels.[11]

At the time of their initial presentation, almost all children have bilateral involvement by arteriography. In the senior author's personal series, however, 16 patients had purely unilateral angiographic and clinical findings at presentation. Over periods ranging from 6 months to 4 years, 9 of these progressed to develop opposite hemisphere symptoms or had development of significant stenoses in previously normal vessels in the opposite hemisphere. Seven patients with unilateral disease, however, did not progress during follow-up periods of up to 7 years after their initial arteriogram and surgery, a finding which again indicates how variable a given moyamoya patient's clinical course can be.

In addition to signs and symptoms, obtaining a careful history is important in patients suspected to have moyamoya. There are a host of prior clinical conditions or syndromes that have been associated with moyamoya.[12] These include prior radiotherapy to the head or neck for optic gliomas, craniopharyngiomas, and pituitary tumors; genetic disorders, such as Down syndrome, neurofibromatosis type I (with or without hypothalamic-optic pathway tumors), large facial hemangiomas, sickle cell anemia, and other hemoglobinopathies; autoimmune disorders such as Graves' disease; congenital cardiac disease; renal artery stenosis; meningeal infections, including tuberculous meningitis; and a host of unique syndromes such as Williams, Alagille, and so on. Table 2 summarizes the clinical associations noted in our recently published series.[10a]

PATHOLOGY AND ETIOLOGY

In moyamoya syndrome, stenosis occurs in the distal intracranial carotid arteries and often involves the proximal anterior and middle cerebral arteries. Pathologic analysis has demonstrated that affected vessels generally do not exhibit arteriosclerotic or inflammatory changes. While the etiology of moyamoya remains unknown, several investigations have made some inroads toward understanding this

Table 2 Associated Conditions, Risk Factors, or Syndromes

Syndrome	Number
No associated conditions (idiopathic)	66
Neurofibromatosis type I	16
Asian	16
Cranial therapeutic radiation	15
(hypothalamic-optic system glioma = 8)	
(craniopharyngioma = 4)	
(medulloblastoma, with Gorlin's syndrome = 1)	
(acute lymphocytic leukemia, intrathecal chemotherapy = 2)	
Down syndrome	10
Congenital cardiac anomaly, previously operated	7
Renal artery stenosis	4
Hemoglobinopathy (sickle cell = 2, ''Bryn Mawr'' = 1)	3
(Other hematologic: spherocytosis = 1, idiopathic thrombocytopenic purpura = 1)	2
Giant cervicofacial hemangiomas	3
Shunted hydrocephalus	3
Idiopathic hypertension requiring medication	3
Hyperthyroidism (with Graves' disease = 1)	2

Other syndromes, 1 patient each: Reyes (remote), Williams, Alagille, cloacal extrophy, renal artery fibromuscular dysplasia, and congenital cytomegalic inclusion virus infection (remote). Two patients had unclassified syndromic presentations. There were 4 African Americans, 2 of whom had sickle cell disease.

disease. Basic fibroblast growth factor has been shown to be elevated in assays of dura and scalp arteries in patients with moyamoya syndrome as well as in the cerebrospinal fluid (CSF) of children with moyamoya syndrome, sampled at the time of revascularization surgery.[13–16] Intracellular adhesion molecules are similarly elevated in the CSF of these patients.[17] These findings suggest that there may be an as-yet-undefined systemic process underlying the vasculopathy in certain patients. Morphometric analyses have demonstrated similar abnormalities in pulmonary, renal, cardiac, and pancreatic arteries in certain patients with moyamoya syndrome.[18] Moreover, the fact that moyamoya syndrome occurs in patients with congenital cardiac and urogenital anomalies, including patients with Down syndrome and in patients with renal artery stenosis, suggests that in these conditions there may be a common genetic alteration in the makeup of vascular structures.[19]

Evidence exists supporting the premise that genetic factors are important in the pathogenesis of this disorder. A familial incidence in Japan of 7 to 12% and a rate of affected first-degree relatives of ~6% in the Children's Hospital, Boston, series substantiate this hypothesis.[20,21] Genetic analysis has suggested associations between moyamoya patients and abnormalities on chromosomes 3, 6, and 17.[22,23] It is hoped that further study of this disorder will lead to improved understanding and, ultimately, better therapy for moyamoya syndrome.

In addition to genetic factors, it appears that environmental influences contribute to the pathogenesis of moyamoya syndrome. The finding that patients can develop this syndrome in a delayed fashion after radiation therapy or after acquiring certain types of infectious illnesses lend credence to the role of nongenetic causes of moyamoya.[19,24–26] In the senior author's personal series, there are two sets of identical twins, only one sibling in each pair having moyamoya syndrome, again suggesting that some type of environmental factor must precipitate the syndrome's clinical emergence in susceptible patients. Ultimately the pathogenesis of moyamoya syndrome may involve both genetic and environmental factors.

NATURAL HISTORY AND PROGNOSIS

The prognosis of moyamoya syndrome is difficult to predict because the natural history of this disorder has not yet been elucidated. The progression of disease can be slow, with rare, intermittent events, or it can be fulminant, with rapid neurologic decline.[10a,12] However, regardless of the course, it seems clear that moyamoya syndrome, both in terms of arteriopathy and clinical symptoms, inevitably progresses in untreated patients.[4]

Overall prognosis of patients with moyamoya syndrome depends on the rapidity and extent of vascular occlusion, the patient's ability to develop effective collateral circulation, the age at onset of symptoms, the severity of presenting neurological deficits and degree of disability, and the extent of infarction seen on computed tomography or magnetic resonance imaging (MRI) studies at the time of initial presentation.[27] In general, neurologic status at time of treatment, more so than age of the patient, predicts long-term outcome.[10a,28]

Importantly, if surgical revascularization is performed prior to disabling infarction in moyamoya syndrome, even if severe angiographic changes are present, the prognosis tends to be excellent. However, if left untreated, both the angiographic process and the clinical syndrome invariably progress, producing clinical deterioration with potentially irreversible neurological deficits over time.[29]

DIAGNOSIS

Moyamoya syndrome should be considered and diagnostic evaluation begun in any child who presents with symptoms of cerebral ischemia (e.g., a TIA manifesting as episodes of hemiparesis,

speech disturbance, sensory impairment, involuntary movement, and/or visual disturbance), especially if the symptoms are precipitated by physical exertion, hyperventilation, or crying. The diagnosis of moyamoya is confirmed by radiographic studies. Signs of moyamoya can be direct, such as the characteristic arterial narrowing and "puff of smoke" collaterals, or they can be indirect, such as evidence of cerebral hypoperfusion or multiple infarcts. Radiographic evaluation of a given patient suspected of having moyamoya usually proceeds through several studies.

The workup of a patient in whom the diagnosis of moyamoya syndrome is suspected typically begins with a head computed tomography (CT) scan. Commonly, small areas of hypodensity suggestive of stroke are observed in cortical watershed zones, basal ganglia, deep white matter, or periventricular regions. Although rare in children, hemorrhage from moyamoya vessels can be readily diagnosed on head CT, with the most common sites of hemorrhage being the basal ganglia, ventricular system, medial temporal lobes, and thalamus.

Patients with these findings on CT are often subsequently evaluated with an MRI/magnetic resonance angiography (MRA). Acute infarcts are well seen using diffusion weighted imaging, chronic infarcts are better delineated with T1 and T2 imaging, and cortical ischemia may be inferred from fluid attenuated inversion recovery (FLAIR) sequences which demonstrate linear high signal following a sulcal pattern, felt to represent slow flow in poorly perfused cortical circulation.[30] Most suggestive of moyamoya on MRI is the finding of diminished flow voids in the internal carotid and middle and anterior cerebral arteries coupled with prominent collateral flow voids in the basal ganglia and thalamus. These imaging findings are virtually diagnostic of moyamoya syndrome.

Because of the excellent diagnostic yield and noninvasive nature of MRI, it has been proposed that MRA be used as the primary diagnostic imaging modality for moyamoya syndrome instead of conventional cerebral angiography.[31–37] While MRA affords the ability to detect stenosis of the major intracranial vessels, visualization of basal moyamoya collateral vessels and smaller vessel occlusions is frequently subject to artifact. Therefore, to confirm the diagnosis of moyamoya syndrome and to visualize the anatomy of the vessels involved and the patterns of flow through the hemispheres, conventional cerebral angiography is typically required.

Angiography should consist of a full four-vessel series, including selective injection of the external carotid systems. The diagnosis of moyamoya is said to require bilateral symmetrical stenosis or occlusion of the terminal portion of the intracranial carotid arteries as well as the presence of dilated collateral vessels that develop at the base of the brain producing the classic "puff of smoke" appearance on angiography. External carotid imaging is essential to identify pre-existing collateral vessels, so that surgery, if performed, will not disrupt them. Aneurysms or arterio venous malformations (AVMs), known to be associated with some cases of moyamoya, can also be best detected by conventional angiography.

The angiographic appearance of moyamoya has been classified into six progressive stages.[4] The first stage is characterized by carotid stenosis without collateral vessels. In the second stage, basal collateral vessels are seen and in the third stage, these vessels become more prominent, often associated clinically with symptomatic presentation of the patients. During the fourth stage, the entire circle of Willis and posterior cerebral arteries become severely stenotic or completely occluded, moyamoya vessels begin to narrow, and extracranial collateral networks begin to form. It is the prominence of these extracranial vessels which is the hallmark of the fifth stage, and they become the only source of blood supply to the brain following complete occlusion of the carotids in the sixth stage.

Other diagnostic studies that can assist in the workup of patients with moyamoya syndrome include electroencephalography (EEG) and cerebral blood flow studies. Specific alterations of EEG recordings are usually observed only in pediatric patients. These include posterior or centrotemporal slowing, a hyperventilation-induced diffuse pattern

of monophasic slow waves (i.e., build-up), and a characteristic "rebuild-up" phenomenon.[38] This "rebuild-up" phenomenon, which occurs after hyperventilation and involves the appearance of characteristic slow waves after disappearance or attenuation of initial slowing, is an EEG finding characteristic of moyamoya syndrome and is observed in roughly 50% of children with this disorder. After ~10 minutes, the rebuild-up resolves and the EEG pattern returns to baseline.

Cerebral blood flow studies, utilizing techniques such as transcranial Doppler ultrasonography, xenon-enhanced CT, positron emission tomography, and single photon emission computed tomography with acetazolamide challenge, also can be helpful in the diagnostic evaluation of patients with moyamoya syndrome as well as assisting in treatment decisions. For example, transcranial Doppler examination provides a noninvasive way to follow changes in blood flow patterns over time in larger cerebral vessels, while xenon CT, positron emission tomography, single photon emission computed tomography can be used both to detect regional perfusion instability prior to treatment and to determine the extent of improvement of functional perfusion after therapy.[39–44]

Although each of these studies has the potential to add information in the diagnosis and management of moyamoya, not all are routinely used at our institution. In our patients, MRI/A and conventional angiography are the standard diagnostic tools utilized for most patients with moyamoya; following surgical treatment, an angiogram and an MRI/MRA are obtained 1 year after operation, and depending on the age of the patient, subsequent yearly MR imaging is prescribed as well.

TREATMENT CONSIDERATIONS

Once a major stroke or hemorrhage has occurred, children with moyamoya syndrome frequently are left with permanent neurologic impairment.[12,45] Therefore, early diagnosis and prompt treatment of this disorder are of utmost importance to prevent additional neurologic deficits. Despite this urgency, there is no agreed-upon method of treatment for patients with this chronic occlusive cerebrovascular disorder. There are reports of some patients who stabilize clinically without intervention, but this typically occurs after they have experienced significant, debilitating neurologic disability.

MEDICAL TREATMENT

Currently, there is no known medical treatment capable of reversing or stabilizing progression of moyamoya syndrome. However, there is support for the use of two classes of medications to slow the progression of the disease: anticoagulants/antiplatelet agents and vasodilators.

The antiplatelet effect of aspirin is useful in moyamoya because some ischemic symptoms appear to occur as a consequence of emboli from microthrombus formation at sites of arterial stenoses.[10a,12,45,46] At The Children's Hospital, Boston, children with moyamoya are treated with lifelong aspirin therapy, with those younger than 6 years of age receiving 81 mg/day and at a variable dose in older children, depending on the presence or absence of symptoms. While anticoagulants like warfarin are rarely used due to the difficulty of maintaining therapeutic levels in children, we have begun to use low-dose low-molecular weight heparin (Lovenox) at 0.5 mg/kg twice a day subcutaneously for selected children, particularly those children who are neurologically unstable and need rapidly reversible anticoagulation prior to procedures such as surgery or angiography that preclude aspirin use. In these cases, the aspirin is held for 10 days prior to the procedure and the heparin is employed as a "bridge" to provide some protection that has a shorter pharmacologic half-life than aspirin. After the procedure is completed, aspirin is usually restarted.

The other medication class that has been useful in the treatment of certain symptoms in

moyamoya syndrome is calcium channel blockers.[45] These drugs may be particularly useful in ameliorating symptoms of intractable headaches or migraines, commonly seen in moyamoya patients, and also seems to be effective in reducing both the frequency and severity of refractory TIA.

SURGICAL TREATMENT

There are several studies in the literature that support a role for surgical management of moyamoya disease, and surgery is generally recommended for the treatment of patients with recurrent or progressive cerebral ischemic events and associated reduced cerebral perfusion reserve. Many different operative techniques have been described, all with the main goal of preventing further ischemic injury by increasing collateral blood flow to hypoperfused areas of cortex, using the external carotid circulation as a donor supply.[10a,12]

Various bypass procedures have been performed in the treatment of moyamoya disease, which can generally be divided into direct and indirect types. Direct anastomosis procedures, most commonly superficial temporal artery (STA) to middle cerebral artery (MCA) bypasses, may achieve instant improvement in focal cerebral perfusion, but these procedures are often technically difficult to perform because small pediatric patients often do not have a large enough donor scalp artery or recipient middle cerebral artery to allow for a anastomosis large enough to supply a significant amount of additional collateral blood supply. Because of proximal stenoses, new blood supply provided to a single MCA branch may not allow wide redistribution of the newly available collateral. Temporary occlusion of a middle cerebral branch during the anastomosis may interfere with leptomeningeal collateral pathways already present and lead to an increased incidence of perioperative stroke.

A variety of indirect anastomotic procedures have been described: encephaloduroarteriosynangiosis (EDAS) whereby the STA is dissected free over a course of several inches and then sutured to the cut edges of the opened dura; encephalomyosynangiosis (EMS) in which the temporalis muscle is dissected and placed onto the surface of the brain to encourage collateral vessel development; and the combination of both, encephalomyoarteriosynangiosis (EMAS).[47–49] There are multiple variations of these procedures, including solely drilling burr holes, without vessel anastomosis,[50,51] and craniotomy with inversion of the dura in hopes of enhancing new dural revascularization of the brain.[52] Cervical sympathectomy and omental transposition or omental pedicle grafting have also been described.[53–63] Increased levels of angiogenic factors, such as basic fibroblast growth factor—which has been shown to be significantly elevated in the CSF of patients with moyamoya syndrome—may contribute to the effectiveness of both indirect (i.e., EDAS, EMS, pial synangiosis, and burr holes) and direct (i.e., STA-MCA bypass) revascularization techniques by enhancing the formation and ingrowth of new blood vessels from extracranial sources.[14,16,64]

Another variation of indirect revascularization is to split the dura to improve collateral development. At surgery, a portion of the dura near the branches of the middle meningeal artery is split into outer and inner layers, and the split surface of the outer layer is placed on the cortical surface. This procedure, combined with a standard EDAS, demonstrated effective cortical revascularization through the dural arteries as well as from the scalp arteries. A histological study of the dura in cases of moyamoya disease showed an increased number of blood vessels in the outer layer.[65] The use of a muscle flap to provide revascularization may lead to brain edema due to mass effect from the muscle pedicle itself. To address this, one group proposed that following EDAMS with dural pedicle insertion, the bone flap be shaved to half of its original thickness and the patient be routinely placed on mannitol postoperatively.[66] Finally, several groups have reported improved results in the use of combined direct and indirect anastamoses.[55,56,67]

At The Children's Hospital, Boston, we utilize a modification of the EDAS procedure termed "pial synangiosis" to treat moyamoya syndrome in both children and adults, which we believe leads to a superb induction of new collateral vessels in the patient with chronic ischemia due to moyamoya.[10a] The technique involves the following steps: (1) a scalp donor artery (most commonly, the posterior branch of the superficial temporal artery) is dissected from distal to proximal along with a cuff of galea and surrounding soft tissue; (2) a large craniotomy is turned in the region that is subjacent to the artery; (3) the dura is opened into at least six flaps to increase the surface area of dura exposed to the pial surface and thereby enhance formation of collateral vessels from the dural vascular supply; (4) the arachnoid is opened widely over the surface of brain exposed by the dural opening (Fig. 1); and (5) the intact donor artery is sutured directly to the pial surface using four to six interrupted 10–0 nylon sutures placed through the donor vessel adventitia and the underlying pia (Figs. 2,3). The bone flap is replaced over a Gelfoam cover of the dura, which is left widely open, and carefully secured to avoid compression of the donor artery. The temporal muscle and skin edges are carefully closed with absorbable sutures to similarly avoid compression of the donor vessel. The rationale behind this procedure is that opening the arachnoid removes a barrier to the ingrowth of new blood vessels into the brain from the sources provided to it; the donor vessel's adventitia is sutured to the pial surface to maintain its contact with the brain in areas where the arachnoid has been cleared.

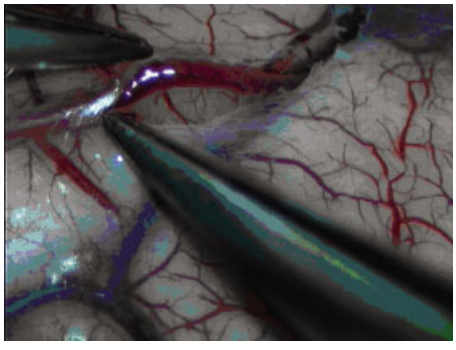

Figure 1 Photomicrograph showing opening of the arachnoid over a cortical vessel using fine forceps.

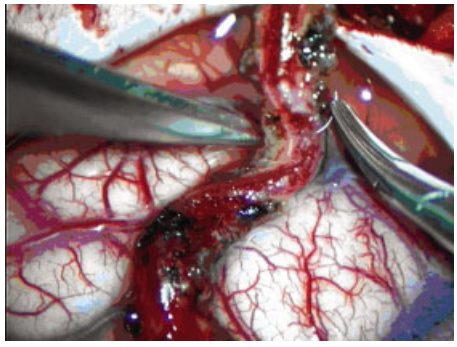

Figure 2 Photomicrograph showing suture (10–0 nylon) passed though adventitia of donor vessel prior to pial stitch.

Once the decision for surgical therapy has been made, several perioperative considerations need to be addressed. In addition to the general issues regarding surgery in children, moyamoya patients are at particular risk of ischemic events in the perioperative period. Crying and hyperventilation, common occurrences in children at times during hospitalization, can lower $PaCO_2$ and induce ischemia secondary to cerebral vasoconstriction. Any techniques to reduce pain—including the use of perioperative sedation, painless wound dressing techniques, and absorbable wound suture closures—helped to reduce the incidence of strokes, TIAs, and length of stay in a recent study.[68] A further perioperative consideration is the use of monitoring, such as intraoperative EEG or near-infrared spectroscopy, used to identify and

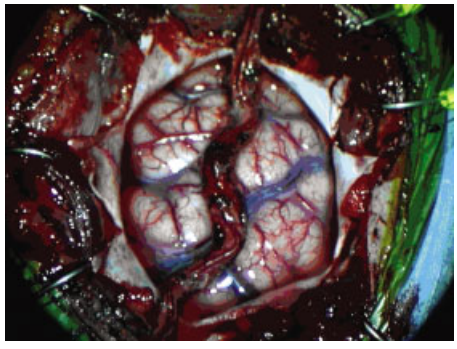

Figure 3 Photograph of donor artery after pial synangiosis showing attachment to cortical surface and wide arachnoidal opening.

ameliorate ischemic events detected while the patient is under general anesthesia.[69,70] At our institution, we have been using full-scalp EEG monitoring during procedures where both hemispheres will be operated on during the same anesthetic. It has helped us identify situations in which continuing with the second side was felt to be unsafe for the patient, but its definitive role is yet to be established. Many EEG changes are observed throughout the procedures that are difficult to interpret. We have noted that the addition of thrombin to our Gelfoam pledget covering the brain at the time of closure is associated with almost invariable EEG slowing, and we have since omitted the use of thrombin at this stage of the operation with a marked reduction in the occurrence of this phenomenon.

Postoperative angiograms are usually obtained 12 months after surgery and typically demonstrate excellent MCA collateralization from both the donor STA and the meningeal arteries. A review of 143 children with moyamoya syndrome treated with pial synangiosis showed marked reductions in their stroke frequency after surgery, especially after the first year postoperatively. In this group, 67% had strokes preoperatively, 7.7% had strokes in the perioperative period, and only 3.2% had strokes after at least 1 year of follow-up. The long-term results are excellent, with a stroke rate of 4.3% (2 patients in a group of 46) in patients with a minimum of 5 years of follow-up.[10a] This work supports the premise that pial synangiosis provides a significant protective effect against new strokes in this patient population.

PERIOPERATIVE AND INTRAOPERATIVE CONSIDERATIONS IN THE SURGICAL MANAGEMENT OF PATIENTS WITH MOYAMOYA SYNDROME

Risks of surgery are more often related to neurologic instability of the patient at the time of surgery and to the risks of anesthesia rather than to actual surgical manipulations. The administration of general anesthesia can result in transient, but significant, physiologic changes which can affect cerebral blood flow. Blood pressure, blood volume, and $PaCO_2$ require careful monitoring because moyamoya patients have a diminished cerebral perfusion reserve and deviation from normal levels can result in stroke.

To reduce the risk of intraoperative and perioperative neurologic morbidity, therefore, meticulous management of the patient is required to avoid hypotension, hypovolemia, hyperthermia, and hypocarbia both intraoperatively as well as perioperatively.[10a] As noted above, intraoperative EEG monitoring with a full array of scalp electrodes can be helpful in the neurologic assessment of patients under general anesthesia. To help prevent hypovolemia during surgery, we admit patients the evening before surgery for aggressive intravenous hydration. Postoperatively, the patients are hydrated with intravenous fluids at one and one-half the normal maintenance rate based on weight for 48 to 72 hours. Aspirin is given on the first postoperative day.

Potential complications associated with surgical treatment of moyamoya syndrome include postoperative stroke, subdural hematoma, both following trauma and spontaneous, and intracerebral hemorrhage. Although wound infection and CSF leak can occur after any cranial procedure, we have only one such a complication in our pial synangiosis series which now numbers over 200 pediatric patients—despite the fact that the dura is left widely open following the synangiosis.

CONCLUSIONS

Moyamoya syndrome is an increasingly recognized entity which is associated with cerebral ischemia. Diagnosis is made on the basis of clinical and radiographic findings, including a characteristic stenosis of the internal carotid arteries in conjunction with

abundant collateral vessel development. Treatment is predicated on revascularization of the ischemic brain, which can be direct (STA-MCA bypass) or indirect (including pial synangiosis). The use of pial synangiosis is a safe, effective, and durable method of cerebral revascularization in moyamoya syndrome and should be considered as a primary treatment for moyamoya, especially in the pediatric population.

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